ABSTRACT

Incontinentia Pigmenti (IP) is a neurocutaneous genodermatosis that segregates as an X-linked dominant disorder with a high probability of prenatal male lethality. A locus in Xq28 containing NF-κB Essential Modulator, a gene product involved in the activation of NF-kB and central to many pro-inflammatory and apoptotic pathways, contains mutations in the majority of cases of IP. Disclosed are methods, compositions and kits directed to a defect in a NF-κB related disease such as IP.

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